Incomplete Kawasaki disease with retropharyngeal space abscess as the main manifestation: a case report and literature review

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Case Report

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Abstract

Background: Kawasaki disease (KD) is a childhood acute self-limiting systemic vasculitis of unknown etiology. The diagnosis of KD is based upon clinician's recognition of a symptom pattern. However, manifestations of KD can be atypical, Deep neck infections (DNIs) is a rare manifestation of KD, which can mislead the clinician and delay diagnosis. The following case report describes a patient with incomplete Kawasaki disease (iKD) whose initial presentation a retropharyngeal abscess.

Case presentation: A child with clinical manifestations of fever, neck pain, cervical lymphadenopathy, high erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) was reported. The anti-infective treatment was not effective. A retropharyngeal space abscess was detected on neck enhanced Computed Tomography (CT). On the third day following admission, the conjunctival hyperemia, red lips, and strawberry tongue appeared gradually. High-dose of intravenous immunoglobulins (IVIG) and oral aspirin were administered with rapid defervescence, fever and neck pain gradually disappeared, and the neck Magnetic Resonance Imaging (MRI) showed that the abscess was completely resolved.

Conclusion: KD with a retropharyngeal space abscess as the first manifestation is rare and may be related to *Streptococcus* or *Staphylococcus aureus* infection. When anti-infective treatment was not effective, it is very important to provide timely diagnosis and treatment in combination with other clinical features of KD.

Background

Kawasaki disease (KD) is the most common cause of childhood acquired heart disease in developed countries. [1] The typical clinical manifestations of KD include fever, pleomorphic rash, nonsuppurative cervical lymphadenopathy, conjunctival injection, strawberry tongue, diffuse injection of the lips and oral mucosa, reddening, and edema of palms and soles at the initial stage, and periungual desquamation at the convalescent stage. However, manifestations of KD can be atypical, which can mislead the clinician and delay diagnosis. For instance, it may only manifest as unexplained fever, redness at the site of BCG inoculation, perianal desquamation, etc., while KD with Deep neck infections (DNIs) as the first manifestation is rare.

Case Presentation

A previously healthy 10-year-old girl presented with fever for 8 days. She had neck pain on the left side with cervical lymphadenopathy, accompanied by sore throat and foreign body sensation in the pharynx, restricted neck movement, accompanied by irritability. Then she went to the otorhinolaryngologic department, physical examination found the pharyngeal cavity was narrow and the posterior pharyngeal wall swelled. Under indirect laryngoscopy, purulent secretions of the epiglottis can be seen. Laboratory findings were as follows: blood routine examination: white blood cell count (WBC): 19.8 $\times 10^9$/L, percentage of neutrophils (N%): 87.4, percentage of lymphocyte (L%): 10.2, hemoglobin (Hb): 137 g/L,

platelets (PLT): 287 $\times 10^9$/L, C-reactive protein (CRP): 54.6 mg/L. The neck computed tomography (CT) showed a left retropharyngeal space abscess. Considering the position of the abscess was deep, and the risk of surgical puncture was high, she was given treatment with cephalosporin antibiotics and ornidazole anti-infective for 6 days, but the fever and neck pain was not relieved, then she transferred to our hospital for treatment.

After admission, physical examination revealed that apart from the swelling and pain of the skin behind the left ear and neck, and cervical lymphadenopathy, the lips diffuse reddening, but there was no strawberry tongue, no abnormal secretions in both ears, no edema of palms and soles, and no periungual desquamation. Repeat blood routine examination: WBC: 20.7 $\times 10^9$/L, N%: 89.5, Hb: 108 g/L, PLT: 321 $\times 10^9$/L, CRP: 80.4 mg/L. Cerebrospinal fluid examination was normal. Ceftriaxone and metronidazole were given to anti-infection treatment. On day 2 of hospitalization, the patient still had recurrent high fever, the thermal spike was not significantly lower than before, irritability, the pain behind the left ear and neck did not relieve, and there was bulbar conjunctival injection and discomfort in both eyes. Vancomycin was added Anti-infection, and tobramycin eye drops are symptomatic. On day 3 of hospitalization, the child's thermal spike was lower than before (38.5℃), and the pain behind the left ear and neck was relieved than before, but the bulbar conjunctival injection and reddening of lips were obvious than before, and strawberry tongue appeared. Laboratory investigations revealed that Serum alanine aminotransferase (ALT): 263 U/L, Aspartate aminotransferase (AST): 62 U/L, erythrocyte sedimentation rate (ESR): 67 mm/h, repeated urination routinely indicated that white blood cells were elevated (5–10 cells/HP), but no urinary irritation symptoms. Echocardiography showed that the left ventricular systolic function and the measured values of left and right coronary arteries and branches were normal, and mitral and tricuspid regurgitation (mild). The neck CT showed a low-density lesion in the left retropharyngeal space, with peripheral enhancement (Fig. 1: arrow) as well as multiple lymph node enlargements in the posterior cervical space. Upon re-evaluation of her clinical manifestations and laboratory findings, the diagnosis of incomplete Kawasaki disease (iKD) was considered, and IVIG at 2 g/kg along with aspirin at 50 mg/kg/day were given. Fever, the left ear and neck pain, and other KD symptoms subsided within 24hours after administration. On day 6 of hospitalization, repeat laboratory investigations revealed blood routine (WBC, CRP), and erythrocyte sedimentation rate was normal. Repeat the neck MRI showed that the abscess was completely resolved, and a low dose of aspirin (5 mg/kg/d) was taken orally.

Discussion And Conclusions

Deep neck infections (DNIs) is a rare manifestation of KD, which include retropharyngeal, parapharyngeal, and peritonsillar abscess.[2] A Population-Based Study among children aged < 12 years showed that of 20,787 patients with Kawasaki disease in the US, 0.6% (130 cases) had DNIs.[3] DNIs often have a rapid onset and can progress to life-threatening complications, such as airway obstruction, jugular vein thrombosis, mediastinal involvement, pericarditis, pneumonia, and arterial erosion.[4] Therefore, timely diagnosis and treatment are necessary. The otolaryngologist may be the initial physician to evaluate a

KD patient with parapharyngeal or retropharyngeal space abscess due to its many head and neck manifestations. Although some typical clinical manifestations of KD have appeared in those patients, but the treatment also delayed due to the lack of knowledge of KD by otolaryngologists.[5, 6]

Many of the KD patients with retropharyngeal space inflammation have not found abscesses or liquefaction during pharynx by fine-needle aspiration.[6–8] It is suggested that cellulitis rather than abscess occurs in KD patients.[9] Yu et al [10] retrospectively analyzed 24 cases of KD patients presenting with retropharyngeal abscess-like edema. None of them were able to fulfill the diagnostic criteria for KD on presentation. They presented as fever with a variety of neck complaints such as neck pain, tender neck mass, torticollis, and limited in range of neck motions. All of those patients were initially managed with empirical antibiotics but were all unresponsive to treatment. As clinical course progressed, more KD signs evolved and fulfilled the complete KD or incomplete KD criteria. One case eventually underwent fine-needle aspiration of the suspicious abscess, subsequent bacterial culture of the aspirated content was negative. After initiation of IVIG, all 24 cases eventually responded satisfactorily to IVIG, this further reinforced the correct diagnosis of KD especially in those cases with incomplete presentation.

However, the causal relationship between the retropharyngeal space inflammation and KD is still difficult to distinguish. Some researchers consider that the retropharyngeal abnormalities in KD are presumably linked to the vasculitis of microvessels that cause oedema and inflammation, which is a non-infectious inflammatory reaction.[11–13] But some researchers consider that the infection of some bacteria such as staphylococcal and streptococcal toxin as superantigens may be involvement in the pathogenesis of KD.[14] Katano et al[15] reported a 10-year-old girl diagnosed with KD, she was subjected to a lymph node biopsy, focal necrosis with inflammatory cell infiltration, including neutrophils and macrophages, was observed in the marginal zone of the cervical lymph node. Streptococcus spp. genome was detected in DNA and RNA samples from a cervical lymph node biopsy specimen. Okada et al[16] reported a 4-year-old boy first presented a peritonsillar and retropharyngeal abscess-like lesion, Surgical tonsillectomy was performed to avoid a risk of mediastinal abscess, but he fulfilled the diagnostic criteria of KD after the operation. Histopathology revealed small lymphatic follicles and neutrophil infiltration in the peritonsillar muscle layer, *Streptococcus α-haemolyticus* were isolated from both throat and aspiration cultures. Choi et al[17] reported a 3-year-old boy with KD with a coexistent parapharyngeal abscess, After treatment with IVIG (2 g/kg/day) and aspirin (80 mg/kg/day), KD symptoms subsided within 24 hours of IVIG infusion, but the painful left neck swelling persisted and severe torticollis developed. He was treated with ceftriaxone, however, there was no improvement of the left cervical lymphadenitis. The neck CT showed a low density lesion with an irregular thick wall in the left lateral node, suggesting an abscess. After incision and drainage of the parapharyngeal abscess, the cervical adenopathy decreased in size and his torticollis began to resolve. A culture of the abscess revealed the growth of *Staphylococcus aureus.*

CT scan was helpful in terms of differentiating deep neck abscesses from cellulitis. Holt et al[18] defined the characteristics of a deep neck abscess as a cystic appearance, low-density CT number, air or fluid at the center of the suspected area, and "rim enhancement". In KD patients, the extent of retropharyngeal phlegmon was typically long-segment with no definite "rim enhancement" collections on CT. [9, 10] So the

inflammation of the retropharyngeal space caused by KD is often manifested as low-density lesions in the retropharyngeal space on CT. In this condition, early suspicion of KD and subsequent satisfactory response to IVIG may avoid the need for antibiotic treatment or invasive procedures such fine needle aspiration or surgical explorations. However, if neck CT showed a retropharyngeal space abscess, it often indicates that there may be a purulent bacterial infection and may be involved in the onset of KD. Our case is similar to the case reported by Choi et al,[17] the neck CT showed tissues "rim enhancement" which suggesting an abscess. The difference is that our case after treatment with IVIG and aspirin, his neck pain and abscess subsided and no need for incision and drainage. The common offending pathogens of retropharyngeal abscess include *Streptococcus*, *Staphylococcus aureus* and epidermidis, with other gram-positive bacteria and anaerobes also seen.[19] Generally, it is not recommended to continue treatment with antibiotics after diagnosis in children with KD, which may aggravate the systemic vascular inflammation. But in our opinion, it is reasonable to managed with empirical antibiotics for KD patient with a retropharyngeal space abscess as the main manifestation.

In summary, we should be alert to the patients with DNIs that show a poor response to initial intravenous antibiotics, as it could be early presentations of KD. KD with deep neck abscess is not an indication for surgery, timely initiation of Kawasaki disease treatment is the key.

Abbreviations

KD
Kawasaki disease
iKD
incomplete Kawasaki disease
DNIs
Deep neck infections
WBC
White blood cell count
N%
Percentage of neutrophils
L%
Percentage of lymphocyte
Hb
Hemoglobin
PLT
Platelets
CRP
C-reactive protein
ESR
erythrocyte sedimentation rate
ALT

Serum alanine aminotransferase
AST
Aspartate aminotransferase
CT
Computed Tomography
MRI
Magnetic Resonance Imaging
IVIG
intravenous immunoglobulins.

Declarations

Ethics approval and consent to participate

This study was approved by the Research Ethics Committee of West China Second University Hospital, Sichuan University.

Consent for publication

Informed consent was obtained from the patient's parents for publication of this case report.

Availability of data and materials

Not applicable.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

JC collected the data, reviewed literature, drafted the manuscript. YW read and approved the final manuscript. All authors read and approved the manuscript.

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Figures

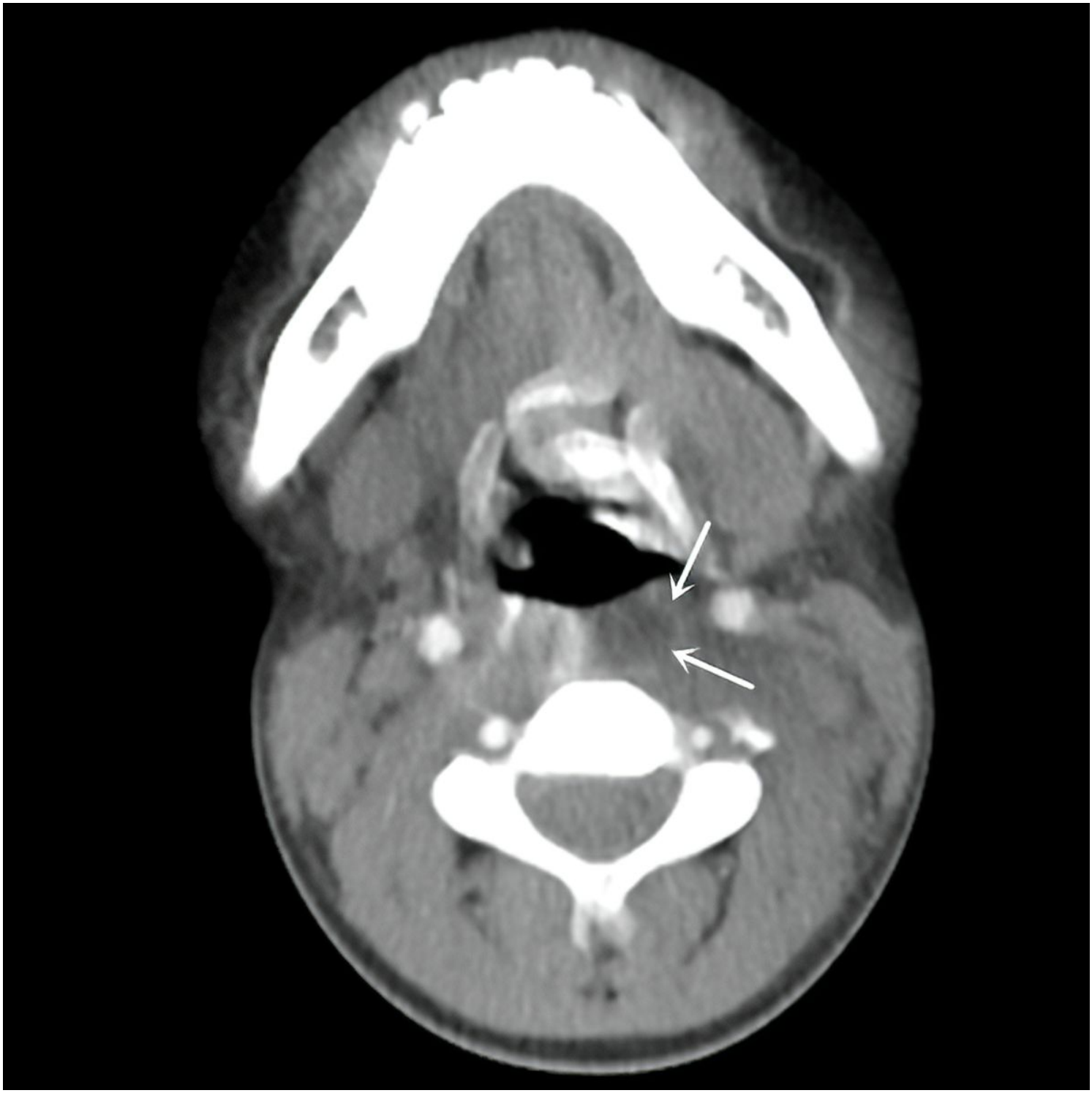

Figure 1

Neck CT scan showed a hypodense lesion in the left retropharyngeal space with peripheral enhancement (arrow).

Supplementary Files

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